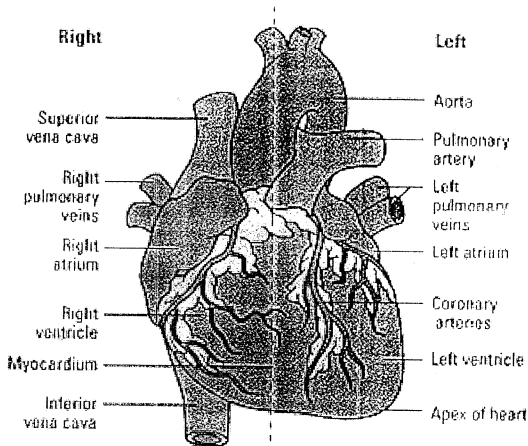


The Heart: Outside



Apex → 27 sec.

77 → 2 sec

77 sec

Fourth space outside middle c.

~
Pituitary

cut

Cardiology

Congenital Heart Diseases

Incidence

- It is the commonest congenital anomalies 30 %.
- This equal 8/ 1000 live births.
- The commonest are VSD, PDA & ASD.

Etiology

- Multifactorial inheritance means that abnormalities of the gene concerned with heart development (+ve family history) on the presence of other environmental factors lead to heart defects, if the environmental hazards occur during 2-9 weeks of pregnancy. *Con'd*
 - The most common cause is idiopathic.
 - Congenital infections e.g. congenital rubella syndrome.
 - Teratogenic effect of drugs e.g. aspirin, lithium, warfarin or irradiation.
 - Associated with chromosomal disorders. Down, Marfan, Turner & Noonan syndromes.
 - Associated with some maternal diseases as DM, PKU, SLE.

Classification

- CHD are classified into acyanotic & cyanotic HD.

Acyanotic	Cyanotic
A. Shunts: - VSD - ASD - PDA B. Obstructive - Aortic stenosis - Pulmonary stenosis C. Malposition - Dextrocardia	A. With oligemic lung: - Fallot's tetralogy - Tricuspid atresia B. With plethoric lung: - Transposition of great vessels - Truncus arteriosus

- All cases with Lt to Rt shunt are considered potentially cyanotic.

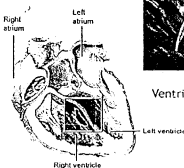
Ventricular Septal Defect (VSD)

Definition

- It is defect in the interventricular sept

Etiology

- Idiopathic type.
- congenital infection: e.g. congenital rubella syndrome
- Teratogenic factors: e.g. drugs during pregnancy as aspirin
- Chromosomal: e.g. Down's syndrome.



Ventricular septal defect

Types**A. According to the site:**

1. Upper $\Rightarrow$ Membranous
2. Lower $\Rightarrow$ Muscular

B. According to the Size:

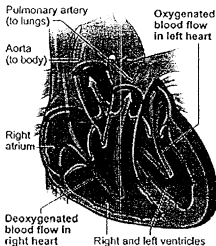
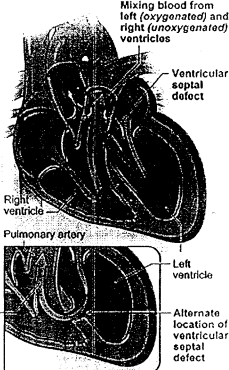
1. Small: $< 0.5\text{cm}$
2. Average: $0.5 - 1\text{cm}$
3. Large: $> 1\text{cm}$

Hemodynamics**1. Small defect:**

- No significant changes & pulmonary hypertension does not occur.
- 75 % will have closure of the defect by 10 years of age (Roger's disease)

2. Large defects:

- The blood From the Lt ventricle is divided into 2 portions one part passes to the aorta and the other one passes to the Rt ventricle $\Rightarrow$ RV++.
- All this blood will pass through the pulmonary artery $\Rightarrow$ Pulmonary plethora $\Rightarrow$ Symptoms of pulmonary congestion.
- Then, pass to the Lt atrium $\Rightarrow$ Lt atrium dilatation.
- Then, to the Lt ventricle $\Rightarrow$ LV dilatation.
- Persistent of pulmonary plethora $\Rightarrow$ P++ $\Rightarrow$ Low COP symptoms.
- Pulmonary hypertension $\Rightarrow$ $\uparrow\uparrow\uparrow$ Rt ventricular pressure $\Rightarrow$ Eisenmenger Syndrome $\Rightarrow$ Persistent cyanosis

A Normal heart**B Hearts with ventricular septal defects**

Down $\Rightarrow$ Common AV canal

Down 13 $\Rightarrow$ Midline defect.

Turner $\Rightarrow$ Conduction of aorta

Marfan $\Rightarrow$ C. aorta incompetence.

Marfan $\Rightarrow$ C. Pulmonary stenosis

Turner $\Rightarrow$ A. arch anomaly.

Alagans $\Rightarrow$ P. stenosis.

Clinical Picture

Symptoms:

1. Asymptomatic: this is the usual presentation in small VSD which is accidentally discovered during routine examination of the heart.
2. Congestive lung symptoms.
3. Palpitation. *the size of woman but give oliguria*
4. Low COP symptoms: when he developed pulmonary hypertension.
5. Potential cyanosis: especially on crying due to $\uparrow\uparrow$ pulmonary pressure $\Rightarrow$ transient reversal of the shunt.
6. Permanent cyanosis: when he developed Eisenminger syndrome.

Signs:

A. Inspection & palpation:

1. Pericardial bulge: in long standing cases.
2. Apex:
 - Site: shifted downward & outward.
 - Size: diffuse.
 - Character: hyper dynamic.
 - Thrill: systolic maximum on the Lt parasternal area.
3. Parasternal pulsations.
4. Epigastric pulsations.
5. Diastolic shock over the pulmonary area in cases complicated by P⁺⁺

B. Percussion: dullness over the pulmonary area in cases complicated by P⁺⁺.

C. Auscultation:

- ❖ S1: normal.
- ❖ S2: $\uparrow\uparrow\uparrow$ in cases complicated by P⁺⁺.
- ❖ Murmur:
 - Timing: Pan systolic.
 - Character: Harsh.
 - Site of maximum intensity: Lt 3rd & 4th parasternal area.
 - Propagation: all over the pericardium.

Complications

1. Repeated chest infections.
2. Infective endocarditis
3. Congestive HF.
4. Eisenminger syndrome $\Rightarrow$ reversal of the shunt due to development of P⁺⁺ $\Rightarrow$ permanent cyanosis & clubbing of the nails.
5. Stunted growth.

Investigations

1. CXR

All chambers of the heart are enlarged except the Rt atrium.

- Large pulmonary arteries
- Pulmonary plethora.

2. ECG $\Rightarrow$ Evidence of Bi-ventricular hypertrophy and Lt atrial dilatation
 - P- Pulmonale with the development of P⁺⁺



3. Echo - Cardiography

Can demonstrate the site, size & direction of flow through the shunt.

4. Cardiac catheterization.**Treatment**A. Medical TTT:

Prophylaxis against infective endocarditis.

B. Surgical TTT: It is indicated in:

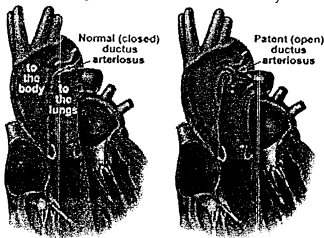
1. Membranous VSD regardless the size.
2. Large muscular VSD.
3. Increasing pulmonary artery pressure.
4. If still present after the age of 5 years.

Roger's disease $\Rightarrow$ small VSD in the muscular part of the septum. There's no hemodynamic disturbance.

- Usually accidentally discovered
- O/E $\Rightarrow$ Harsh loud ejection systolic murmur maximum over the 3rd & 4th Lt parasternal area
- Spontaneous Closure is the rule

PDA**Definition**

It is persistence of the fetal communication between the main pulmonary artery & the aortic arch after the origin of the Lt subclavian artery.

**Mechanism of closure**1. Functional closure

- At birth $\Rightarrow$ blood passes from the aorta to the pulmonary through DA, this blood is oxygenated $\Rightarrow$ stimulation of local axon reflex $\Rightarrow$ inhibition of PGE2 release from the endothelial cells of DA $\Rightarrow$ contraction of the wall of the ductus This occurs within 10-15 hrs of birth

2. Later, clots form $\Rightarrow$ fibrosis $\Rightarrow$ obliterative closure.

Spontaneous closure may occur up to 3 months after birth of premature.

From Above

1. Maternal PGE2 $\Rightarrow$ PDA
2. Neonatal anoxia $\Rightarrow$ PDA
3. Anti-prostaglandins $\Rightarrow$ closure of the ductus
4. Congenital rubella $\Rightarrow$ Proliferation of endothelial $\Rightarrow$ $\uparrow$ PGE2 $\Rightarrow$ PDA

Hemodynamics

1. The pulmonary receives blood from the aorta & RV $\Rightarrow$ Lung plethora $\Rightarrow$ LA++ $\Rightarrow$ LV++
2. After sometime $\Rightarrow$ P++ occur $\Rightarrow$ Reversal of the shunt $\Rightarrow$ Eisenminger syndrome $\Rightarrow$ Persistent cyanosis in the lower half of the body "Differential Cyanosis"

Clinical PictureSymptoms:

1. Asymptomatic: This is the usual presentation in small PDA.
2. Congestive lung symptoms.
3. Palpitation.
4. Low COP symptoms: in the lower limbs only.
5. Differential cyanosis in the lower limbs only: when he developed Eisenminger syndrome.

Signs:

- General signs:
 - 1) Water hammer pulse.
 - 2) Cyanosis & clubbing in both lower limbs when he developed Eisenminger syndrome
- Local signs:
 - 1) Inspection & palpation:
 - Apex:
 - Site: shifted downward & outward.
 - Size: localized
 - Character: hyper dynamic.
 - No parasternal pulsations.
 - No epigastric pulsations.
 - Diastolic shock over the pulmonary area in cases complicated by P++.
 - Thrill: can be felt below the left infra-clavicular area.
 - 2) Percussion:
 - Dullness over the pulmonary area in cases complicated by P++.
 - 3) Auscultation:
 - S1: normal.
 - S2: in cases complicated by P++
 - Murmur:
 - Timing: continues.
 - Character: machinery.
 - Site of maximum intensity: below the left clavicle.
 - Propagation: to the pulmonary area & back.



Complications:

1. Repeated chest infections.
2. Infective endocarditis.
3. Congestive HF.
4. **Eisenminger syndrome** $\Rightarrow$ reversal of the shunt due to development of P++ $\Rightarrow$ permanent cyanosis & clubbing of both lower limbs.
5. Stunted growth mainly in the lower segment.

Investigations:

1. CXR

- All chambers of the left side of heart are enlarged.
- Large pulmonary arteries
- Pulmonary plethora.

2. ECG

- Evidence of Lt-ventricular and Lt atrial dilatation
- P- Pulmonale with the development of P⁺⁺.

3. Echo-Cardiography

- Can demonstrate the size & direction of flow through the shunt.

4. Cardiac catheterization

Treatment:

A. Medical:

- Prophylaxis against infective endocarditis.
- If the condition diagnosed early anti-prostaglandin as Indomethacin can be given.

B. Surgical TTT: It is indicated as soon as possible

ASD

Definition

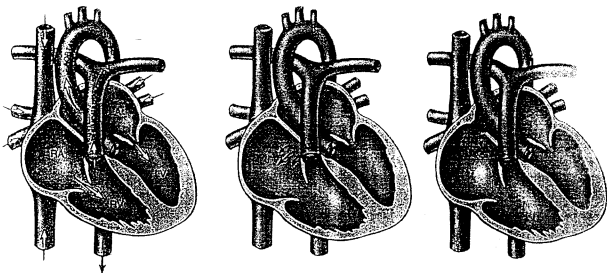
- It is defect in the inter-atrial septum

Etiology

- As VSD

Types

1. Ostium primum $\Rightarrow$ defect in the lower part of the septum "less common"
2. Ostium secundum $\Rightarrow$ defect in the upper part of the septum "more common"
3. Patent foramen ovale
4. Endocardial Cushion defect "common A-V canal" Ostium primum defect + High VSD + TI $\pm$ MI
 the common congenital heart disease in Down's syndrome



Hemodynamics

1. Lt atrial pressure is more than the RT atrium So part of blood is shunted from LA to RA which receives blood from the superior & inferior vena-cava $\Rightarrow$ RA ++.
2. This big amount of blood goes to the RV $\Rightarrow$ RV++ $\Rightarrow$ to the pulmonary A. $\Rightarrow$ enlargement of the pulmonary artery, $\Rightarrow$ lung plethora $\Rightarrow$ repeated chest infection.
3. Then to LA where part of blood is shunted to RA & the rest goes to LV $\Rightarrow$ Aorta.

CP

1. Small defect usually asymptomatic
2. Dyspnea & recurrent chest infection "due to pulmonary plethora".
3. Growth retardation
4. O/E
 - Rt ventricular enlargement.
 - Wide, Fixed splitting of the 2nd heart sound on the pulmonary area.

N.B. Passage of blood through the ASD produces no murmurs. But, soft ejection systolic murmur may be heard over the pulmonary area due to function P.S.

Investigations

1. CXR:

- All Chambers of the heart are enlarged except the LV
- Large pulmonary arteries & plethora.

2. ECG:

- RV⁺⁺
- Rt bundle branch block (RBBB)

2. Echo-cardiology & cardiac catheterization $\Rightarrow$ diagnostic

Complications

1. Recurrent chest infections.
2. H.F. "Rt. ventricular"
3. Infective endocarditis is rare due to low pressure gradient.
4. Eisenminger syndrome " uncommon & late"
5. Lutembacher syndrome $\Rightarrow$ ASD + rheumatic Ms.

Treatment

Surgical closure of large defects

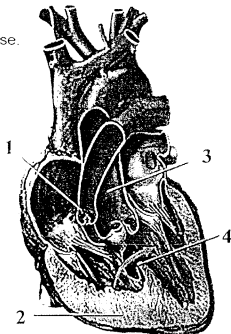
Fallot's tetralogy

Definition

- It is the most common congenital cyanotic heart disease.
- It consists of tetrad
 1. Infundibular "subvalvular" PS
 2. RV hypertrophy
 3. Over-riding aorta
 4. Big membranous VSD.

Hemodynamics

- The blood in the RV will pass through 2 ways.
 1. Small amount passes through the narrow pulmonary to the lung $\Rightarrow$ lung oligemia. This small amount will be oxygenated & pass to LA $\Rightarrow$ LV $\Rightarrow$ aorta.
 2. Larger amount will pass non oxygenated to the overriding aorta $\Rightarrow$ cyanosis.
- The RV enlarges but not markedly because the blood has 2 ways to leave.



Clinical Picture

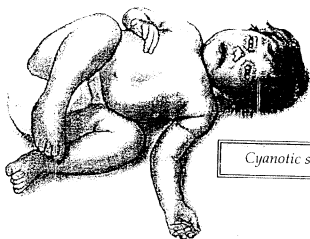
1. Central Cyanosis: appearing shortly after birth --- why?
2. Clubbing of fingers $\Rightarrow$ usually drum stick.
3. Squatting position: Preferred by the patient because.
 - a. Kinking of femoral & popliteal arteries $\Rightarrow$ $\uparrow\uparrow$ resistance to blood going through the aorta & so forces more blood to pass to the lungs.
 - b. Blood is squeezed from L.L. & splanchnic area, so more blood goes to the brain.



4. Cyanotic spills

Due to spasm of the infundibulum forcing all blood in RV to the aorta un氧xygenated $\Rightarrow$ cerebral hypoxia $\Rightarrow$ syncope & cyanosis became more deep $\Rightarrow$ convulsions.

5. Stunted growth



Cyanotic spell

O/E

1. No pulsation or dullness over the pulmonary area.
2. The 2nd sound over the pulmonary area is:
 - a. Single: aortic component only.
 - b. Accentuated: due to $\uparrow\uparrow$ aortic blood flow:
3. Harsh ejection systolic murmur maximum over the pulmonary area "PS".
 - a. Inversely proportionate to severity of PS
 - b. It is $\Rightarrow$ uncommonly accompanied with thrill.

Complications

1. Cyanotic spills
2. Pulmonary T.B.
3. Polycythemia $\Rightarrow$ $\uparrow\uparrow$ thrombosis.
4. Cerebral thrombosis.
5. Brain abscess $\rightarrow$ why?
6. Infective endocarditis & HF are rare.
7. Growth retardation & delayed puberty.

Investigations

1. CXR

a. Coeur en sabot:

- Enlarged aorta.
- Small pulmonary artery (exaggerated waist)
- RV enlargement.

b. Oligemic lung fields.

2. ECG $\Rightarrow$ slight RV hypertrophy.

3. Echo-cardiography & cardiac catheterization.



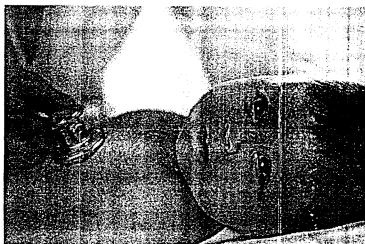
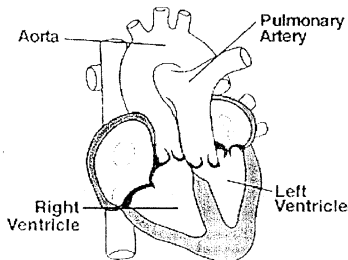
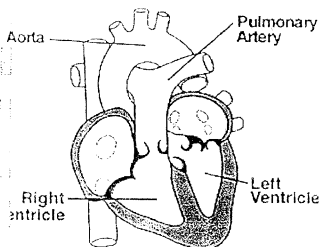
Treatment

1. Protect against infective endocarditis.
2. Propranolol 1mg / Kg / 6hrs $\Rightarrow$ to prevent cyanotic spells.
3. Cyanotic spells.
 - a. Put the patient in squatting position.
 - b. O_2 for correction of anoxia.
 - c. $NaHCO_3$.
 - d. Inderal IV 0.1-0.2 mg / kg $\Rightarrow$ relax the infandibular spasm.
 - e. Morphine (0.2 mg/kg) $\Rightarrow$ in resistant cases.
4. Iron therapy.
5. Surgical:
 - a. Palliative $\Rightarrow$ pulmonary systemic anastomosis e.g. Blalock-Taussing operation between Lt pulmonary & Lt subclavian artery.
 - b. Brock operation = infandibular resection.
 - c. Correction through open heart surgery.



Transposition of Great Arteries

- In this condition the aorta arises from the Rt ventricle & the pulmonary artery from the Lt ventricle. So, non oxygenated blood enters the systemic circulation.
- Survival depends on the mixing between systemic & pulmonary circulations commonly via patent foramen ovale or ASD & PDA.
- In early infancy, the patient's condition may deteriorate when the DA closes.
- **C/P:** The baby is healthy at birth but becomes persistently cyanosed within the 1st day of birth. Later signs of P++ develop, and if neglected HF will occur.
- **X-ray:** Egg on side appearance of cardiac shadow.



Rheumatic Fever

Etiology

It is an auto-immune disease related to throat infection by streptococcus hemolyticus through one of the following mechanisms:

1. Fixation of streptococcal toxins into tissue proteins rendering them antigenic, and stimulate antibody formation against them.
2. Antibodies formed against streptococcal antigen are immunologically identical to antigenicity of the heart.

Predisposing Factors

1. Age: 5-15 years (rare below 3 years or above 25 years).
2. Sex: equal (Chorea common in females)
3. Familial tendency.
4. Season: cold months.
5. Over crowding & bad hygienic conditions.
6. Recurrent streptococcal infection.

Clinical Picture

Modified Jonne's criteria

A. Major	B. Minor
1. Arthritis	1. Fever
2. Carditis.	2. History of rheumatic fever or rheumatic heart disease.
3. Chorea.	3. Arthralgia
4. Erythema marginatum	4. +ve acute phase reactants
5. Subcutaneous nodule	5. Prolonged P-R interval

C. Evidence of streptococcal infection

1. Recent scarlet fever
2. +ve throat cultures
3. ↑ ASOT & other streptococcal antibodies

Rheumatic fever can be diagnosed if there are

- ** 2 major + evidence of streptococcal infection.
- ** 1 major + 2 minor + evidence of streptococcal infection

N.B :

- 1) If arthritis is present ⇒ arthralgia is not a minor.
- 2) If carditis is present ⇒ prolonged P-R interval is not a minor.
- 3) Rheumatic chorea may not be accompanied by other manifestations or evidence of streptococcal infection !

1. Carditis:

A. Pericarditis "Dry"	B. Myocarditis	C. Endocarditis
1. Stitching pain 2. Pericardial rub	1. Tachycardia out of fever 2. Gallop rhythm 3. Muffled heart sounds. 4. Dilatation of the ring ⇒ incompetent valves 5. HF in severe cases. 6. ECG: P-R interval 7. CXR: dilated heart	1. Edema of the mitral valve ⇒ MS ⇒ transient mid-diastolic murmur <u>Carey comb's</u> murmur. 2. Destruction & deformity ⇒ MI ± AI 3. On previous Rh heart ⇒ change of character of previous murmur or appearance of new murmur

2. Arthritis:

- Usually large joints e.g. knees, ankle hip, shoulder, elbow or wrist.
- Involved joints become ⇒ red, hot, swollen, tender + loss of function.
- Fleeting arthritis.
- Leaving no residual damage or deformity
- Responds rapidly to salicylates

3. Chorea:

Due to involvement of basal ganglia "Caudate nucleus"

- Common in females around age of puberty.
- It may present alone without other major or minor manifestations.
- Character: Involuntary, semi-purposeful, sudden jerky movements of all limbs, face & tongue.
- Increased by emotions & excitement & disappear during sleep.
- Hypotonia & hyporeflexia are usually associated.
- Emotional instability is usually present.
- Course: self limited or prolonged

4. Subcutaneous nodules:

- Painless, freely mobile under the skin.
- Site ⇒ most commonly over the extensor surfaces & bony prominence as elbows, knees, chin of tibia & mastoid process.

5. Erythema marginatum

- Rings of erythema, with clear center & irregular border.
- Site ⇒ on the trunk.
- They are not painful, not itchy & not scaly.
- They extend to coalesce together with central fading of erythema.

Investigations:1. Acute phase reactants.

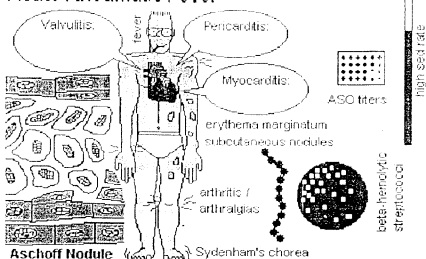
- Leucocytosis.
- ↑↑ ESR
- C-Reactive protein +ve.

2. ECG ⇒ prolonged P-R interval & tachycardia.3. Evidence of previous streptococcal infection

- ↑↑ ASOT > 250 Todd's unit.
- ↑↑ Other streptococcal antibodies.

4. X-ray chest & heart. ⇒ Cardiomegaly.5. Throat culture ⇒ group A, beta hemolytic streptococci may still be present.6. Echo-cardiography: ⇒ Evidence of pancarditis & valvular affection.

Acute Rheumatic Fever



Treatment

A. Prophylaxis**a. Prevention of 1st attack:**

1. Proper hygiene to prevent streptococcal infections. This includes good housing, avoiding over crowding, good nutrition, & avoid contact with infected children.
2. Early detection & proper TTT of streptococcal throat infections.
 - The course of TTT should be at least 10 days, using procaine penicillin or erythromycin.
 - A single IM injection of long acting penicillin is advised by some authors.

b. Prevention of 2nd attack of Rh. fever:Prophylaxis against streptococcal infections for:

- At least 5 years after the last attack.
- Up to 25 years if there's cardiac lesion.
- Some authors recommend continuation for Life.

The drugs used are:

- Long acting penicillin 1.2 million units / 2 weeks.
- Oral penicillin V (Ospen) $\Rightarrow$ 250,000 IU once or twice / day
- Erythromycin 250 mg twice / day.
- Sulfadiazine 0.5-1 gm once / day.

c. Prevention of infective endocarditis:

1. Procedures which predispose to bacteremia. e.g. tooth extraction, tonsillectomy, catheterization $\Rightarrow$ should be done under an umbrella of antibiotics in all patients with history of rheumatic or congenital heart diseases. $\Rightarrow$ Crystalline penicillin 250,000 IU + procaine penicillin 600,000 IU by IMI one hour before operation. $\Rightarrow$ Then $\Rightarrow$ penicillin V 250,000 — 500,000 IU / 8hrs for 2 days oral.

Recently

- Oral amoxicillin 50 mg/kg 1hr before the procedure & 25 mg/kg 6hrs after the initial dose.
- If there's penicillin allergy $\Rightarrow$ Erythromycin 20 mg/kg 2 hrs before the procedure & 10mg / kg 6hrs after..

GIT or UT procedures $\Rightarrow$ Gentamycin 2mg / kg or streptomycin 20 mg / kg is given with the initial injection.

Recently: $\Rightarrow$ parenteral ampicillin 50 mg / kg + Gentamycin 2 mg/kg are given 1/2 hr before the procedure.

* If there's penicillin allergy $\Rightarrow$ Vancomycin 20mg / kg slow IV.

2. Early tt for upper respiratory infections & other septic focus to prevent bacteremia.
3. Proper dental care & oral hygiene.

B. Curative TTT

1. Anti-inflammatory drugs:

- a. Salicylates: 100-120 mg / kg / D for 1-2 weeks after clinical improvement or normal ESR.

Indication $\Rightarrow$ Arthritis without carditis.

- b. Prednisone: 2mg/kg/D "max 60mg" for 4-6 weeks or until ESR is normal $\Rightarrow$ then $\Rightarrow$ gradual tapering over 2 weeks under an umbrella of salicylates to prevent rebound. Salicylates are continued for 4 weeks.

Indication $\Rightarrow$ Carditis.

2. Antibiotics

To eradicate streptococci $\Rightarrow$ procaine penicillin 400,000 IU / D for 10 days IMI or erythromycin 20-40 mg/kg/day in 4 divided doses for 10 days. Then followed by long acting penicillin.

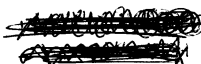
3. Bed rest

Until ESR returns normal especially if carditis is present.

4. TTT of heart failure if present:

- Low salt diet.
- Lasix: 2-3 mg / kg / D
- K-syrup
- Digitalis: digitalization 0.05-0.07 mg/kg as follow
 - 1/2 the dose is given immediately
 - 1/4 after 8 hrs.
 - 1/4 after 8 hrs.Then maintenance 0.01 mg / kg / D

4. TTT of rheumatic chorea:

- Self limited disease, but usually needs supportive care, cheerful environment & understanding attendants.
 - Phenobarbitone: 3-5 mg / kg / day. or
 - Chlorpromazine: 1 mg / kg / day. or
 - Diazepam: 0.2 mg / kg / day. or
 - Haloperidol: 1-5 mg total dose / day.
- 

Infective Endocarditis

Infection of endocardium may be due to bacteria, rickettsiae or fungus.

Classification

A. Sub-acute form "SBE"

- Caused by low virulent organism.
- Affects diseased heart.

B. Acute form "ABE"

- Caused by highly virulent organism.
- Affects normal & diseased heart.

Infective endocarditis is an infection of the heart chambers or valves



Etiology

There is combination of 2 factors:

Bacteremia	Underlying cardiac lesion
The causative organism 1. Streptococcal viridians "50%" 2. Streptococcal fecalis 3. Staphylococcus aureus $\Rightarrow$ may affect normal heart. 4. G-ve bacteria common in neonates & IV drug abusers. The most common is H-influenza. The Root of infection 1. Dental extraction 2. GIT & urinary procedure. 3. Cardiac surgery.	Congenital: $\Rightarrow$ VSD, PDA & coarctation of the aorta Rare $\Rightarrow$ ASD why? Rheumatic $\Rightarrow$ MI, AS & AI. Rare $\Rightarrow$ MS $\Rightarrow$ why? Prosthetic valves

Fungal endocarditis:

It is due to Candida albicans, aspergillus species etc.

Population affected are:

- Post cardiac surgery.
- Narcotics.
- Immune-compromised patients.
- Neonates with prolonged use of IV catheters.

Pathogenesis

2 factors are needed for the formation of vegetations.

High pressure source	Narrow orifice
So, it is more common in Lt sided heart lesion & rare in: • Heart failure. • MS • ASD	So It is more common in small than big VSD

Clinical Picture

Triad

- A. Embolic manifestations.
- B. Toxemic manifestations.
- C. Underlying cardiac lesion.

A. Toxemic Manifestations

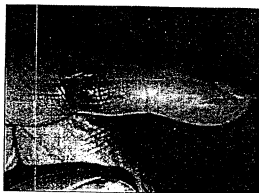
- o **Fever** $\Rightarrow$ continuous or intermittent fever of long duration
 It may be high grade but the most common low grade fever.

- o Anorexia, general weakness & loss of weight
- o Marked pallor & toxic facies.
- o Eyes:

- Roth spots.
- Subconjunctival hge.

- o Hands

- Pale clubbing
- Splinter hge in the nail beds
- Ossler's nodules: Small, tender, intra-cutaneous nodules in pulps of the fingers, opposite the heads of metacarpal bones
- Jane way nodules $\Rightarrow$ small painless, blue on the palms & soles.



These lesions represent vasculitis produced by circulating antigen antibody complexes.

- o Spleen

Slightly enlarged, soft & tender

B. Underlying cardiac lesion

- o Change in the character of the already present murmur
- o Development of new murmur.
- o Rupture cusps or chorda tendinae $\Rightarrow$ loud musical murmur "Seagull murmur".
- o Endocarditis may precipitates HF

C. Embolic manifestations

The vegetations are easily detached $\Rightarrow$ septic or aseptic emboli which may lead to:

1. Infarction: if end artery is affected.
2. Ischemia: if non end artery is affected.
3. Mycotic aneurysm due to low grade infected emboli.
4. Renal affection:
 - Flea bitten kidney $\Rightarrow$ Microscopic hematuria
 - Acute glomerulonephritis $\Rightarrow$ RBCs cast
 - Infarction $\Rightarrow$ profuse hematuria.

Complications

1. Embolization.
2. Renal affection $\Rightarrow$ renal failure.
3. HF due to extensive endocarditis.

Investigations

1. Blood culture & Sensitivity.

3 samples for aerobic, anaerobic bacteria & fungi.

2. CBC: Leukocytosis & Anemia.

3. ESR

4. Urine analysis: Hematuria.

5. Echo-cardiography: To see the vegetations.

Treatment

A. Prevention $\Rightarrow$ see rheumatic fever.

B. Curative

Antibiotic TTT for at least 4-6 weeks according to culture & sensitivity

a. It must be bactericidal

b. Big doses are needed by IVI.

- A combination of penicillin G 200,000 IU / kg / day IV & gentamycin 2-4 mg / kg / day IV.
- If the organism is resistant to penicillin give nafcillin 200 mg / kg / day IV or Vancomycin 40 mg / kg day IV or 3rd generation cephalosporin's.

Heart Failure

Definition

- Failure of the heart to supply the tissues with their needs of blood & oxygen.

Etiology

- HF occurs as a result of
 - Pressure overload
 - Volume over load
 - Myocardial disease.



1. Congenital heart diseases

- Large left to right shunt : VSD, ASD & PDA.
- Right & left side obstructive lesions: PS, AS & coarctation of the aorta.
- Congenital valve defect: congenital MI.
- "N.B." It is very rare in F4.

2. Acquired valvular lesions: usually 2ry to rheumatic endocarditis e.g. MS, MI AS, AI.

3. Carditis

- Rheumatic $\Rightarrow$ most common after 5Y.
- Viral myocarditis $\Rightarrow$ Coxsackie B virus "commonest"
- Toxic myocarditis $\Rightarrow$ Pneumonia, Septicemia, Diphtheria & Typhoid.
- Infective endocarditis.

4. Arrhythmias: either tachyarrhythmia (e.g. supraventricular tachycardia & atrial fibrillation) or bradyarrhythmia (e.g. complete heart block).

5. High cardiac output $\Rightarrow$ severe anemia, thyrotoxicosis, A-V fistula.

6. Cardiomyopathy & endocardial fibroelastosis.

7. Hypertension & pulmonary embolism.

8. Cor pulmonale as in cases of cystic fibrosis & bronchopulmonary dysplasia.

9. Fluid overload particularly in the newborn.

Clinical Picture

Depends on the severity of failure.

In infants:

Symptoms:

- Tachypnea & feeding difficulty.
- Poor weight gain.
- Excessive perspiration.
- Irritability & weak cry.

Signs:

- Signs of respiratory distress.
- Tender hepatomegaly.
- Generalized edema more in the eye lids.
- Heart: tachycardia, gallop rhythm + original cardiac lesion.

In children:

1. Low C/O symptoms & signs.

- Easy fatigability.
- Lack of concentration.
- Syncopal attacks.

- Oliguria.
- Intermittent claudication.
- Rapid weak pulse.
- Pale cold extremities.

2. Congestive lung symptoms & signs (in LVF only).

- Cough & expectorations.
- Dyspnea, orthopnea & paroxysmal nocturnal dyspnea.
- Hemoptysis.
- Repeated chest infections.
- O/E: Basal consonating crepitation on the basal part of the lung,

3. Manifestations of systemic venous congestion (in RVF).

- Dyspepsia & vomiting.
- Pain in the right hypochondria.
- Edema in both lower limbs.
- O/E:
 - a. Congested pulsating neck veins.
 - b. Enlarged tender liver.
 - c. Pitting edema in both LL.
 - d. Generalized anasarca in severe cases.

4. Gallop rhythm & findings of the original lesion on auscultation.

Investigations

For diagnosis of underlying cardiac lesion & assessment of cardiac functions.

1. CXR:
 - Cardiomegaly.
 - ↑↑ Pulmonary vascularity in shunts.
 - Pulmonary edema in severe cases: fluffy pulmonary markings of venous congestion & acute pulmonary edema.
2. ECG: helps in diagnosis of specific chamber enlargement.
3. Echocardiography: helps in assessment of myocardial function & diagnosis of underlying lesion.
4. Arterial blood gases.
5. Electrolytes especially Na & K.

Treatment

I. General:

- a. Complete rest in bed: in semi-sitting position.
- b. Salt free diet.
- c. O₂ inhalation.
- d. Treatment of underlying-etiology.

II. Diuretics:

- Furosemide (Lasix): 1-2 mg/kg dose IV or 1-4 mg / kg / day oral. + K syrup. (mechanism of action: loop diuretics ⇒ ↓ volume overload ⇒ ↓ work of the heart, ↓ edema & congestion.)
- Spironolactone (Aldactone): 2-3 mg / kg / day oral. (K-sparing).
- Chlorthalidate: 20-40 mg / kg / day.
- Mechanism of action: ↓↓↓ volume over-load → ↓↓↓ work of the heart.

IV. Digitalis: see t t t of rheumatic fever.

Mechanism of action:

- +ve inotropic.
- Slow AV conduction.

V. Vasodilators:

- Na nitroprusside: 0.5-8ug/kg/min IVD in ICU. ($\downarrow\downarrow$ pre & after load).
- Captopril: 0.5-6 mg / kg / day oral (ACE inhibitor $\rightarrow \downarrow\downarrow$ aldosterone $\rightarrow \downarrow\downarrow$ after-load).
- Hydralazine: 0.5 mg / kg IV or oral 0.5-0.75 mg / Kg / D. ($\downarrow\downarrow\downarrow$ after-load).

VI. Cardiac stimulants:

- Isoproterenol: 0.01-0.05 ug /kg / min IVD in ICU only. (β adrenergic effect & improve myocardial contractility & $\downarrow$ after-load).
- Dopamine: 2-20 ug / kg / min IVD in ICU only (as isoproterenol + renal vasodilator).
- Dobutamine: 2-20 ug / kg / min IVD in ICU only.

Mechanism of action: β adrenergic effect $\rightarrow \uparrow\uparrow\uparrow$ myocardial contractility.

VII. Treatment of the underlying cause.

Primary cardiomyopathy

It is a heart muscle disease in the absence of CHD, hypertension, acquired valve lesions, abnormal coronaries, infection or other systemic disease.

Types

1. Hypertrophic cardiomyopathy (AD).

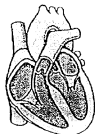
- There is left ventricular hypertrophy associated with hypertrophy of the IVS $\rightarrow$ obstruction of outflow of LV $\rightarrow$ dyspnea, PND, chest pain, syncope, palpitations & sudden death.
- TTT: β -blockers & Ca channel blockers.

2. Dilated cardiomyopathy (commonest) (multifactorial? viral).

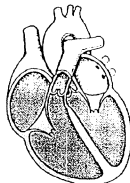
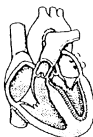
- There is poor contracting myocardium with low COP & CHF.
- TTT: as HF.

3. Restrictive cardiomyopathy.

- There is poor ventricular compliance & inadequate filling.
- It has poor prognosis.



Normal

Dilated
CardiomyopathyHypertrophic
CardiomyopathyRestrictive
Cardiomyopathy

Secondary Cardiomyopathy

I. Heredo-familial:

1. Duchene myopathy.
2. Becker's myopathy.
3. Friedreich's ataxia.

II. Infections:

1. Virus: Coxsackie's A & B, adenovirus.
2. Rickettsiae.
3. Bacteria: diphtheria, TB & typhoid.
4. Parasitic: Toxoplasmosis.
5. Fungi.

III. Metabolic, nutritional & Endocrinal:

1. Beriberi.
2. KWO.
3. MPS.
4. Hypo & hyperthyroidism.
5. Infant of diabetic mother.

IV. Connective tissue, granulomatous & infiltrative:

1. SLE.
2. JRA.
3. Rheumatic fever.
4. Sarcoidosis.
5. Leukemia.

V. Drugs & toxins:

1. Adriamycin.
2. Hemosidrosis.
3. Irradiation.

VI. Coronary artery diseases:

1. Congenital.
2. Kawasaki.

VII. Others:

1. Anemia.
2. Sickle cell anemia.

		VSD	F4	ASD
Symptoms		1- Asymptomatic 2- Congestive lung symptoms 3- Palpation 4- Low CO symptoms 5- Cyanosis on crying (+/-)	1- Cyanosis: starts shortly after birth 2- Clubbing 3- Squatting position 4- Cyanotic spells	1- Symptomatic 2- Congestive lung symptoms 3- Palpitation 4- Cyanosis after reversal of shunt
Inspection and palpation		1. Pericardial bulge. 2. Apex: - Site: shifted down and outward - Size: diffuse - Character: hyper dynamic - Thrill: systolic 3. Lt. para-sternal pulsations. 4. Epigastric pulsations. 5. Diastolic shock over the pulmonary area in case complicated by P++	1. Pericardial bulge. 2. Apex: - Site: may be shifted outwards. - Size: may be diffuse. - Character: normal 3. Lt. parasternal pulsations (+) 4. Epigastric pulsations (+) 5. No thrill pulsations over the pulmonary area.	1. Pericardial bulge. 2. Apex: - Site: shifted outwards. - Size: diffuse. - Character: hyper dynamic. 3. Lt. and Rt. Parasternal pulsations. 4. Epigastric pulsations. 5. Diastolic shock: over the pulmonary area in cases complicated by P++.
Signs	Percussion	Dullness over the pulmonary area in cases complicated by P++	The pulmonary area is resonant	Dullness over the pulmonary area in cases complicated by P++
	Auscultation	1- S1: normal 2- S2: accentuated in cases complicated by P++ 3- Harsh, pan-systolic murmur maximum over the Lt. 3 rd & 4 th para-sternal area propagated all over the pericardium.	1. S1: normal 2. S2: single and accentuated. 3. Harsh, ejection systolic murmur, maximum over the pulmonary area, propagated to the Lt. parasternal area.	1. S1: normal. 2. S2: wide, fixed splitting. 3. Soft, ejection systolic murmur, maximum over the pulmonary area, may be propagated to the Lt. Parasternal area (functional PS)

Complications	1. Repeated chest infections 2. Congestive heart failure. 3. Infective endocarditis 4. Isenminger syndrome 5. Stunted growth	1. Cyanotic spells. 2. Pulmonary TB. 3. Polycythemia → thrombosis 4. Cerebral thrombosis 5. Brain abscess 6. Infective endocarditis & HF. 7. Stunted growth	1. Repeated chest infections. 2. Congestive HF. 3. Infective endocarditis 4. Isenmenger syndrome 5. Stunted growth.
Investigations	1. CXR. 2. ECG. 3. Echocardiography. 4. Cardiac catheterization.		
Medical Treatment	Prophylaxis against SBE.	1. Prophylaxis against SBE. 2. Propranolol: 1mg/kg as prophylaxis against cyanotic spells. 3. Cyanotic spells: - a- Put the patient in squatting position. - b- O₂ therapy. - c- NaHCO₃ - d- Propranolol IVI - e- Morphine in resistant cases.	Prophylaxis against SBE.
Surgical	1- Membranous regardless the size. 2- Large muscular. 3- Increasing pulmonary artery pressure. 4- After the age of 5 years.	- A- Palliative: 1- Blalock-Taussing. 2- Brock - B- Total correction .	Surgical closure of large defect

		PDA	MS	MI
Symptoms		1. Asymptomatic 2. Low CO symptoms (in LL) 3. Congestive lung symptoms 4. Palpitation. 5. Differential cyanosis (in LL) on reversal of shunt.	1. Asymptomatic in mild cases. 2. Congestive lung symptoms. 3. Low CO symptoms (hypertensive type) 4. Symptoms of systemic venous congestion in cases complicated by RVF.	1- Asymptomatic 2- Palpitation 3- Low CO symptoms 4- Congestive lung symptoms.
Signs	Inspection and palpation	1- Apex: - Site: shifted downwards and outwards. - Size: localized. - Character: hyper dynamic 2- Diastolic shock over pulmonary area in cases complicated by P++ 3- Continuous thrill over the Lt. infra-clavicular area.	A- General: 1. Absent in mild cases. 2. Fine basal crepitations in congestive MS. 3. Malar flush in hypertensive MS. 4. Congested neck veins, enlarged tender liver in stage IV. B- Local: 1- Apex: - Site: may be shifted outwards. - Size: diffuse. - Character: slapping. - Diastolic thrill 2- Parasternal pulsations 3- Epigastric pulsations 4- Diastolic shock over the pulmonary area in cases complicated by P++.	A- General: As in MS except malar flush. B- Local: 1- Apex: - Site: shifted downward and outward - Size: localized. - Character: hyper dynamic. - Systolic thrill: 2- Diastolic shock over the pulmonary area in cases complicated by P++.
	Percussion	Dullness over the pulmonary area in cases complicated by P++	Dullness over the pulmonary area in cases complicated by P++	Dullness over the pulmonary area in cases complicated by P++

	Auscultation	1- S1: normal 2- S2: accentuated in case complicated by P++. 3- Continuous machinery murmur, maximum over the Lt. infra-clavicular area, propagated to the pulmonary area.	1- S1: accentuated. 2- S2: accentuated in cases complicated by P++. 3- Mid-diastolic rumbling murmur, maximum over the apex and may be propagated to the inner side of the apex.	1- S1: muffled. 2- S2: accentuated in cases complicated by P++ 3- Soft, pan-systolic murmur, maximum over the apex, propagated to the axilla.
	Complications	1. Repeated chest infections 2. Congestive heart failure. 3. Infective endocarditis 4. Isenminger syndrome 5. Stunted growth	A- Valve: 1- RH-activity. 2- SBE. 3- Calcifications B- Lt. atrium: 1- AF. 2- Pressure manifestations. 3- Thrombo-embolism. C- Lung: 1- Repeated chest infection. 2- Hemoptysis. 3- Pulmonary infarction. D- RVF. 1. CXR. 2. ECG.	As MS
	Investigations	1. CXR. 2. ECG. 3. Echocardiography. 4. Cardiac catheterization	1. CXR. 2. ECG. 3. Echocardiography.	As MS
Treatment	Medical	1- Prophylaxis against SBE. 2- Endomethacin early in the neonatal period.	1- Prophylaxis against SBE. 2- Prophylaxis against rheumatic fever.	As MS
	Surgical	Surgical closure even if small	1- Dilatation in pure non-calcified MS. 2- Valve replacement if there is calcifications or MI.	Valve replacement

		AS	AI
Symptoms		1- Asymptomatic in mild cases. 2- Low CO symptoms. 3- Anginal pain. 4- Congestive lung symptoms if complicated by LVE.	As AS + palpitation
Signs	General	1- Plateau pulse. 2- Systolic thrill over the carotid arteries.	1- Water hammer pulse. 2- De-Musse sign. 3- Corrigan's signs. 4- Prominent capillary pulsations. 5- Pistol shot.
	Inspection and palpation	1. Apex: - Site: shifted downward and outward. - Size: localized. - Character: heavy sustained.	<u>Apex:</u> - Site: shifted downward and outward. - Size: localized. - Character: hyper dynamic. - Diastolic thrill.
	Percussion	Dullness on the 1 st aortic area due to post-stenotic dilatation.	Non-specific
	Auscultation	1. S1: normal. 2. S2: muffled. 3. Harsh ejection systolic murmur, maximum over the 1 st aortic area, propagated to the apex and neck.	1. S1: normal. 2. S2: muffled. 3. Soft, early diastolic murmur, maximum over the 2 nd aortic while the patient is leaning forward.
Complications		As valve complications of MS + myocardial infarction	As valve complications of MS + LVF
Investigations		As MS	As MS
Treatment	Medical	As MS + propranolol for IHSS	As MS + TTT of syphilitic AI
	Surgical	1- Dilatation in pure non-calcified AS. 2- Valve replacement if there is calcifications or AT. 3- Resection for IHSS.	Valve replacement